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Letter to the Editor

Reply-Letter to the Editor-Is liver steatosis diagnostic of non-alcoholic fatty liver disease in patients with hereditary fructose intolerance?

This letter is in response to the letter by Dr. Giuseppe Maggiore et al. entitled *Is liver steatosis diagnostic of non-alcoholic fatty liver disease in patients with hereditary fructose intolerance?*

In their letter, Dr. Maggiore et al. comment that non-alcoholic fatty liver disease (NAFLD) found in patients with hereditary fructose intolerance (HFI) is surprisingly unrelated to obesity, overweight or insulin resistance. This is the novel point of our results and we understand that this may create controversy. In the SSIEM 2018 (Annual Symposium of the Society for the Study of Inborn Errors of Metabolism, Athens, Greece, 4–7 September 2018) it was also presented as an oral communication that adult HFI patients on a fructose-restricted diet accumulate more hepatic fat without any sign of hepatic fibrosis [1].

As we stated in our conclusion, currently there are no data on the physiopathological mechanisms that produce fatty liver in patients with HFI but there are some mechanisms which may be implicated or affected. We are studying some of them in our current funded research projects, such as the beta oxidation of fatty acids [2–5].

We also highlight, based on our results and previous studies, the importance of lipidomic and methylation analyses for understanding the mechanisms that produce fatty liver in these patients as it has been observed that changes in the methylation index are directly associated with a deregulation of the export of hepatic lipids mediated by very low density lipoproteins (VLDL) [6].

To sum up, we believe that it is necessary to explore several mechanisms such as beta oxidation and methylation which could explain the development of NAFLD not related to obesity or insulin resistance.

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